

An Uncommon Presentation in Behçet's Disease: Recovered Descending-Colon Perforation with Conservative Medical Therapy: Case Report

Behçet Hastalarında Nadir Bir Vaka: Konservatif Medikal Tedavi ile Düzelen İnen Kolon Perforasyonu

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ABSTRACT Behçet's disease is a multisystem disorder. Intestinal involvement is common in patients from the Far East, but is rare among those from the Middle East. The most frequent area of involvement is the ileocecal region. Although intestinal perforations of Behçet's disease are most frequently seen in the cecum, perforations of the sigmoid and descending colon have also been reported in the literature. A 45-year-old man with Behçet's disease admitted to our hospital with abdominal pain and active lower gastrointestinal bleeding. A colonoscopic examination revealed multiple perforated ulcers in the descending colon. He was followed-up conservatively, and no surgical intervention was required. Descending-colon perforation is a rare complication of Behçet's disease. Our patient recovered with conservative medical treatment, without surgery. Therefore, this case suggests that medical treatment with close follow-up may be useful in the management of intestinal perforations in Behçet's disease.

Key Words: Behçet syndrome; intestinal perforation

ÖZET Behçet hastalığı multisistemik bir bozukluktur. İntestinal tutulum uzak doğudaki hastalarda sıklıkla ancak orta doğuda nadirdir. En sık tutulum yeri ileoçekal bölgedir. Behçet hastalığında intestinal perforasyonlar çekumda sık görülse de, literatürde sigmoid ve inen kolonda da perforasyonlar bildirilmiştir. Behçet hastalığı olan 45 yaşında bir erkek hasta hastanemize karın ağrısı ve aktif alt gastrointestinal kanama ile başvurdu. Kolonoskopide çok sayıda perfore ülser görülmüştür. Konservatif tedavi ile cerrahi işleme gerek kalmadan izlenmiştir. İnen kolon perforasyonu Behçet hastalığının nadir bir komplikasyonudur. Hastamız cerrahi tedaviye gerek kalmadan konservatif medikal tedaviyle düzelmiştir. Bu nedenle, bu olgu yakın takiple medikal tedavinin Behçet hastalığında intestinal perforasyonun tedavisinde faydalı olabileceğini düşündürmektedir.

Anahtar Kelimeler: Behçet sendromu; intestinal perforasyon

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The highest incidence of Behçet's disease is in the Middle East, the Mediterranean basin, and the Far East. Some of the disease manifestations show regional differences. Intestinal involvement is common in patients from the Far East, but is rare among those from the Middle East,¹ and is characterized by mucosal ulcers. The ileocecal region is the most commonly affected site in the gastrointestinal tract. These intestinal mucosal ulcerations are deep, with a tendency to perforation and penetration.²

The intestinal perforations in Behçet's disease are most commonly seen in the cecum and terminal ileum.³⁻⁷ However, sigmoid⁸ and descending-colon perforations⁹ have also been reported in the literature. We

present a case who developed multiple perforations in the descending colon due to Behçet's disease, because descending-colon perforation is a rare complication of this disease.

CASE REPORT

A 45-year-old man admitted to our hospital with abdominal pain and active lower gastrointestinal bleeding. He had a past history of Behçet's disease which had been diagnosed two years earlier. Previous medical history revealed administration of colchicine 1 mg/day. On a physical examination, he had diffuse abdominal tenderness, especially in the epigastrium. The results of laboratory tests were as follows: white blood cells 25800/mm³, sedimentation rate 70 mm/h, and C-reactive protein 95 mg/dL. Abdominal ultrasound revealed fluid between the intestinal segments. A colonoscopic examination revealed multiple perforated ulcers in the descending colon (Figure 1). Therefore, we promptly discontinued colonoscopic examination. The patient was then consulted with surgeons. On the first day of follow-up in the intensive-care unit, the oral colchicine dose was increased to 1.5 mg/day. In addition, treatment was combined with oral prednisolone 48 mg/day, sulfasalazine 1500 mg/day, and azathioprine 150 mg/day, intravenous ceftriaxone 2 g/day, and ornidazole 1500 mg/day. On the first day of follow-up, his discomfort and white blood cell count (15200/mm³) had clearly decreased. Therefore, he was treated conservatively, and no surgical intervention was required. A clinical recovery was observed with this therapy, and his lower gastrointestinal bleeding stopped. The patient was discharged on the 10th day of hospitalization. His prednisolone dose was gradually reduced to 8 mg per day.

The patient's laboratory test results after 45 days were as follow: white blood cells 8600/mm³, sedimentation rate 10 mm/h, and C-reactive protein 3 mg/dL. He had no discomfort.

DISCUSSION

Behçet's disease is a multisystem disorder characterized by vasculitis, first described by Hulusi Behçet in 1937. It consists of a triad of recurrent ulcers of the oral and genital mucosa, with relap-

sing uveitis.¹⁰ The frequency of gastrointestinal lesions in Behçet's disease is 1-30%.¹ Gastrointestinal involvement is reported in about one-third of patients from Japan, but is relatively rare among patients from the Mediterranean basin. The most frequent area of involvement is the ileocecal region (75%).¹¹ Intestinal perforations in Behçet's disease are seen commonly in the cecum. Perforations at sites in the large bowel other than the cecum are very rare in Behçet's disease in the Mediterranean basin.^{7,8} Therefore, we report a case who developed multiple perforations in the descending colon in Behçet's disease.

The intestinal lesions of Behçet's disease are manifested as mucosal inflammation or ischemia/infarction secondary to small and large vessel involvement, respectively, and range from nonspecific colitis to diffuse ulcers.⁸ The intestinal ulcers in this disease are prone to hemorrhage, fistulization, penetration, and perforation.¹² It is well known that intestinal perforation is a life-threatening complication and an indication for surgery. Surgical treatment has been performed in all cases of perforation in Behçet's disease reported in the literature. Turan et al. reported a patient with sigmoid-colon perforation,⁸ and Arhan et al. reported a patient with descending-colon perforation⁹ in Behçet's disease. In these cases, surgery was undertaken, as in other cases of intestinal perforation. In our patient, we detected multiple perforated ulcers in the descending colon with colonoscopy. Laboratory findings supported the diagnosis of perforation and peritonitis. The patient was consulted with surgeons and prednisolone, sulfasalazine, ornidazole, and azathioprine treatments were administered. The patient was admitted to the intensive-care unit. We decided to treat him conservatively, because his abdominal pain and white blood cell count decreased in the first 24 hours. The clinical findings of perforation decreased dramatically without surgical intervention on the second day of treatment in the intensive-care unit. In literature, no reports up to date reported perforations healed with conservative medical treatment in Behçet's disease. Although the medical treatment of intestinal Behçet's disease remains controversial,



FIGURE 1: Colonoscopy reveals ulcers in the descending colon (arrows).

successful medical treatments such as tumor necrosis factor-alpha inhibitor (infliximab)¹³ and thalidomide³ have been reported in the management of recurrent and perforating intestinal ulcers. The postoperative recurrence rate for intestinal complications in Behçet's disease is very high (40-87.5%).^{2,12} The rate of patients requiring reoperation is 37.5-47%. In the light of these data, close follow-up with medical treatment may help to avoid surgery and its complications.

In summary, descending-colon perforation is a rare manifestation of Behçet's disease. Our patient recovered with conservative medical treatment alone, without surgery. Therefore, this case suggests that close follow-up with medical treatment could be helpful in the management of intestinal perforation in Behçet's disease.

REFERENCES

1. Yazici H, Fresko I, Yurdakul S. Behçet's syndrome: disease manifestations, management, and advances in treatment. *Nat Clin Pract Rheumatol* 2007;3(3):148-55.
2. Kasahara Y, Tanaka S, Nishino M, Umemura H, Shiraha S, Kuyama T. Intestinal involvement in Behçet's disease: review of 136 surgical cases in the Japanese literature. *Dis Colon Rectum* 1981;24(2):103-6.
3. Sayarlioglu M, Kotan MC, Topcu N, Bayram I, Arslanturk H, Gul A. Treatment of recurrent perforating intestinal ulcers with thalidomide in Behçet's disease. *Ann Pharmacother* 2004; 38(5):808-11.
4. Griffin JW Jr, Harrison HB, Tedesco FJ, Mills LR 4th. Behçet's disease with multiple sites of gastrointestinal involvement. *South Med J* 1982;75(11):1405-8.
5. Goldstein SJ, Crooks DJ. Colitis in Behçet's syndrome. Two new cases. *Radiology* 1978; 128(2):321-3.
6. Nilsen KH, Jones SM, Shorey BA. Behçet's syndrome with perforations of the colon. *Postgrad Med J* 1977;53(616):108-10.
7. Chou SJ, Chen VT, Jan HC, Lou MA, Liu YM. Intestinal perforations in Behçet's disease. *J Gastrointest Surg* 2007;11(4):508-14.
8. Turan M, Sen M, Koyuncu A, Aydin C, Arici S. Sigmoid colon perforation as an unusual complication of Behçet's syndrome: report of a case. *Surg Today* 2003;33(5): 383-6.
9. Arhan M, Ibiş M, Koklu S, Ozin Y, Oymaci E. Behçet's disease complicated with descending colon perforation. *Dig Surg* 2005;22(5): 381.
10. Ustun C. A famous Turkish dermatologist, Dr. Hulusi Behçet. *Eur J Dermatol* 2002;12(5): 469-70.
11. Yurdakul S, Tüzüner N, Yurdakul I, Hamuryudan V, Yazici H. Gastrointestinal involvement in Behçet's syndrome: a controlled study. *Ann Rheum Dis* 1996;55(3):208-10.
12. Isik B, Ara C, Kirimlioglu H, Sogutlu G, Yilmaz M, Yilmaz S, et al. Single or multiple perforations with varying locations as a complication of intestinal Behçet's disease: report of three cases. *Scand J Gastroenterol* 2005;40(5):599-603.
13. Byeon JS, Choi EK, Heo NY, Hong SC, Myung SJ, Yang SK, et al. Antitumor necrosis factor-alpha therapy for early postoperative recurrence of gastrointestinal Behçet's disease: report of a case. *Dis Colon Rectum* 2007; 50(5):672-6.